THE RETROGRADE ATROPHY OF THE PYRAMIDAL TRACTS.[1]

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Introduction—The retrograde atrophy of medullated nerve fibers is a centripetal, ascending process, slowly extending from the seat of lesion toward the trophic center.

It appears only after a considerable time, and may occur in the peripheral nerves, as well as the fiber systems of the brain and cord.

It is essentially a slow atrophic process, quite different in character from the acute changes observed in secondary degenerations and the so-called retrograde degenerations.

It must not be confused with the atrophy of the second order. This is a gradual wasting of a system of fibers otherwise intact, because of the functional inactivity into which it has been thrown by an injury to the contiguous neurones of similar function. This form rarely occurs excepting from injuries in early life, a common example of which is the hemiatrophy of the cord following amputation of an extremity.

The retrograde atrophy of the pyramidal tracts at the first glance resembles the usual descending degenerations. On closer scrutiny certain differences become apparent.

The sclerosis is less compact, and contains in its meshes innumerable minute medullated nerve fibers, with attenuated myelin sheaths, giving to the field a somewhat "peppered" appearance.

The chief histological alterations are in the myelin sheath, which is atrophic, fragmented, or absent, as the case may be. The axis cylinders are well preserved, although slightly swollen and staining faintly. The retrograde process extends, as a rule, only a few inches above the lesion, and in none of the recorded cases could be followed above the decussation of the pyramids.

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It is also interesting to note that the pathological changes in the nerve fibers are in a more active stage at the upper limit of the atrophy, i. e., furthest removed from the initial lesion, showing the gradual progression and manner of extension.

The condition is met with but rarely, and is very inconstant in the time required for its development. The number of recorded observations is small.

Michaud[1] described as early as 1871 a degeneration of the pyramidal tracts above a transverse lesion of the cord.

Sottas[2] (1893), however. was the first to describe in detail the special features of the retrograde atrophy of the pyramidal tracts.

Other observations are those of Williamson[3] (1893), Raymond[4] (1894), Gombault and Phillippe[5] (1894), Dercum and Spiller[6] (1896), Dejerine and Sottas[7] (1896), Durante[8] (1898), and Spiller[9] (1898).

In most of these cases the underlying pathological condition was one of many years duration; the syringomyelia, syphilitic patches of sclerosis and myelitis. I can find no record of any case in which the atrophy was apparent before the lapse of two years from the date of the initial lesion.

Gombault and Phillippe noted the retrograde atrophy after two years, and Williamson after two years and four months.

I recently examined the cord from a case of total transverse myelitis of acute onset, situated in the mid-dorsal region, in which the process had been stationary for two years, without finding the faintest indication of a retrograde change.

Report of Case—The case was one of slow compression of the spinal cord extending over a period of at least ten years. In addition to the usual secondary degenerations the retrograde atrophy of the pyramidal tracts was very evident, and could be traced to the decussation of the pyramids, and to a less degree through the medulla and pons. For a complete clinical and pathological record of the case consult the *Medical Record*, June 13, 1903, article entitled "On neurofibromatosis," by Fraenkel and Hunt. The essentials of the case, especially those facts bearing on the present study, are as follows:

A young married woman, aged 25, developed symptoms of a progressive compression of the spinal cord referable to a lesion in the lower cervical region. The onset was most insidious and the progress an extremely slow one.

Owing to the absence of pains and irritative root symptoms no operative procedure was undertaken, although the question was frequently raised and discussed. At an examination made in 1897 by Dr. Joseph Fraenkel, the motor and sensory paralysis was found to be complete, corresponding in level to the seventh and eighth cervical segments of the cord.

There was a total spastic paraplegia with flexion contractures, complete anesthesia to the clavicles, including a strip along the ulnar side of each arm. The intrinsic muscles of the hands, including some of the forearm group, were atrophic, and presented fibrillary twitchings.

This condition remained practically stationary until death, which occurred in the winter of 1902. She had been a patient in the Montefiore Home for ten years, during which time the cord had been undergoing gradual compression. For five years the trans-section of the cord had been practically complete.

In addition there was noted in the subcutaneous tissue of the right abdominal wall a large, firm, nodular tumor (plexiform neuroma), the true significance of which was not recognized until autopsy.

The post-mortem examination furnished an immediate explanation for the peculiar insidiousness and chronicity of the process. Beneath the laminæ of the 7 and 8 c and I D vertebræ was situated a dense fibrous tumor, enclosed by the dura mater. Under the tumor, which was one and one-half inches long, the cord was reduced to a mere ribbon-like remnant, gradually tapering above and below to the normal configuration.

The growth, a neuro-fibroma, and evidently originating in one of the spinal nerve roots, is to be regarded as an extreme local manifestation of a general neuro-fibromatosis, which was exquisitely represented in the plexuses and nerve trunks of the peripheral nerves and the anterior and posterior nerve roots of the cervical region of the cord.

In the light of the pathological findings the plexiform neuroma of the abdominal wall assumes a much greater importance than was attached to it during life, and should have suggested the possible nature of the compression.

The atrophic strand, representing the last remnant of the cord, was composed merely of connective tissue and dense glia, with the calcareous remains of blood vessels and ganglion cells. A few medullated nerve fibers were still demonstrable on the periphery.

Below the lesion the sharply demarcated degeneration of the direct and crossed pyramidal tracts could be followed to their termination.

Above the compressed area, in addition to the usual secondary degenerations of the long ascending tracts, the pyramidal tracts, both direct and crossed, appeared sclerosed.

This degeneration of the pyramidal tracts was so distinct as to first suggest a second lesion at a higher level.

A careful examination of the brain and of the cord above the compression, showing the integrity of these parts, a retrograde atrophy was accepted in explanation.

In order to determine the nature and extent of the atrophy various levels of the cord, medulla, pons and crura were studied by the Van Gieson and Weigert methods.

Microscopical Examination—By the Weigert method the atrophy of the pyramidal tracts above the compressed level is distinct and well defined, and may be traced with ease to the decussation of the pyramids. The semi-lunar areas occupied by the direct pyramidal tracts stand out with especial clearness. Figs. 1, 2 and 3. The distinctness of the field of atrophy of the crossed pyramidal tracts is somewhat masked by the adjacent ascending degenerations and the network of the formatio reticularis of the cervical region of the cord.

On comparison with the descending degenerations (see Fig. 4) the retrograde atrophy is not so sharply defined nor so uniformly white, due to the presence of numerous fine medullated nerve fibers.

All evidence of the retrograde change does not cease, however, at the decussation of the pyramids. The pyramids of the medulla are a trifle smaller and are distinctly paler than normal. At the decussation this pallor is confined to the anterolateral aspect of the pyramid, whereas at a higher level (olive) it is more apparent in the dorsal area. In the pale areas the glia tissue is increased. Many of the pyramidal bundles in the pons are paler and stand out with less distinctness than in normal control preparations.

In that portion of the crura cerebri occupied by the pyramidal tracts there is no visible sign of the slightest change. The histological changes are best studied in the direct pyramidal tracts. The sclerosis following the retrograde change is less intense, and the glia network is not so dense and coarse as in that of the ordinary secondary degeneration. Within the sclerosed area are scattered numerous naked axis cylinders, staining faintly, some of which are moderately swollen. Others are enclosed by a very attenuated myelin sheath. Occasional spaces are seen, containing bare axis cylinders or with the myelin sheath fragmented and broken up.

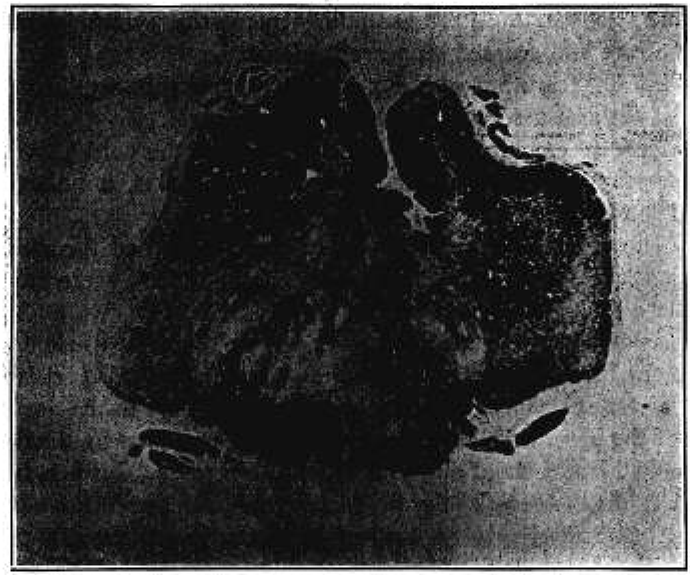

Fig. I. Wiegert-Pal method. Section of cervical cord just above the compressed level, showing retrograde atrophy of the direct and crossed pyramidal tracts.

The paracentral lobules of the Rolandic area were examined by the Nissl method. The large pyramidal cells of this region (cells of Betz) showed no definite pathological changes.

Remarks—The law of Waller not only demands the degeneration of the peripheral portion of the neuraxone, which has been separated from the cell body, but it demands as well the integrity of the central portion of the nerve fiber still in communication with the trophic center.

In recent years considerable controversy has attended the degeneration changes occurring in the central stump of an injured nerve fiber, the so-called retrograde degeneration.

Fig. II. Section from mid-cervical region showing retrograde atrophy of the direct and crossed pyramidal tracts.

Experimental and pathological studies have shown that not infrequently an acute degeneration in the central stump occurs, similar histologically to that described by Waller. It is, therefore, held by some observers that the old conception requires modification.

The consensus of opinion at the present time regards these inconstant and incomplete retrograde degenerations as nothing but descending changes obeying the Wallerian law, and referable to a concomitant disease of the cell body. Among the causes affecting the cell may be mentioned pressure, toxines, a failure of the cell body to undergo reparative changes after the well-known reaction at distance (dislocation of the nucleus and chromatolysis). An ascending neuritis has also been adduced in explanation.

Slow retrograde changes in the nature of an atrophy do occur, however, in the central stump after injury or disease of the nerve fibers of the central and peripheral nervous systems, and is to be distinguished from the mis-called retrograde degeneration. Such atrophy is more pronounced the younger the subject and the nearer the lesion to the trophic center.

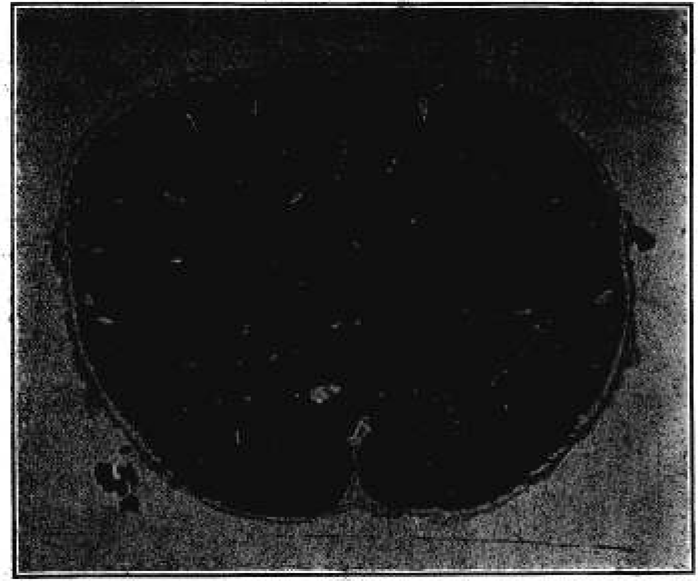

Fig. III. Cervical segment. Retrograde atrophy of direct and crossed pyramidal tracts.

In very young subjects, or the newborn, the central stump may disappear entirely, constituting Gudden's degeneration. The atrophic process in such cases may also extend to the contiguous neurone of similar function, the so-called atrophy of the second order, or the tertiary atrophy, as it is sometimes termed.

In later life the neurones having attained their full development and a greater physiological and anatomical stability, the atrophic changes in the central stump are comparatively slight, and the tertiary atrophy is not apparent.

The retrograde atrophy of the pyramidal tracts is of this nature, and receives its most satisfactory explanation in the prolonged disease and the consequent functional inactivity and nutritional changes which this would entail.

The strictly systematic character of the change, its appearance only after years of disuse. and the comparatively slight histological changes with preservation of the axis cylinders, allow of no other interpretation. The slow extension of the process from the seat of lesion toward the cell is also in harmony with other facts which point to the more distal portion of the neurone as the more vulnerable.

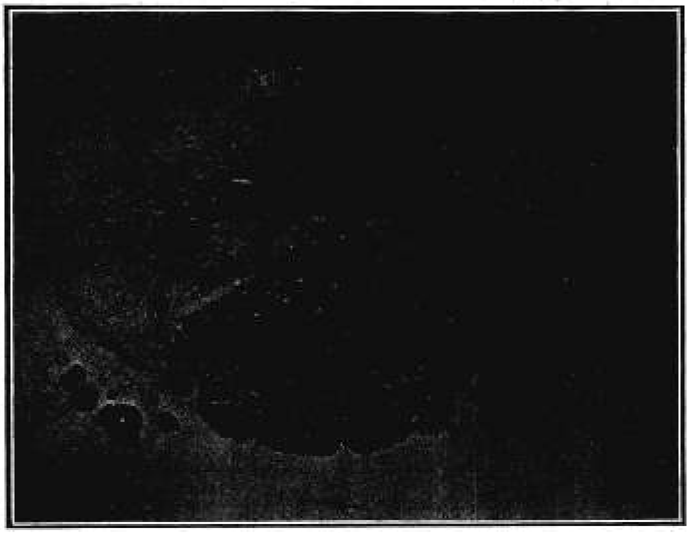

Fig. IV. Wiegert-Pal method. Mid-dorsal region. Descending degeneration of the direct and crossed pyramidal tracts.

This form of atrophy is inconstant, and varies in the time required for its appearance. Although many old cord lesions are subjected to study it is comparatively a rare finding.

This would appear to indicate a variation in the resistance and vital activity in the tracts of different individuals. A fact which has received confirmation under other conditions.

The comparative freedom of the sensory tracts from this form of atrophy is also to be emphasized.

The case which I describe does not differ essentially from those already recorded, except that very distinct evidences of atrophy were present in the pyramidal tracts of the medulla and pons, although milder in degree than that observed below the decussation.

The gradual fading away of the process in ascending is due partly to its nature and partly to the constant addition and admixture of normal nerve fibers from the higher levels.

It is also interesting to note the integrity of the cells of Betz. They were not only normal in configuration and constituents, but were remarkably free from any undue pigmentary change.

The secondary ascending sclerosis, which follows the atrophy, appears to be a simple replacement proliferation not compromising the adjacent normal fibers, which have entered the cord at higher levels.

This freedom from secondary implication of the normal fibers in the sclerosed area is to be inferred from the absence of spastic phenomena in the upper extremities, the nerve supply of which is above the lesion yet coursing through the sclerosis. This also favors the theory of the simple secondary nature of the atrophy.

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